

# Omalizumab Trial in a Patient with Malignant Idiopathic Anaphylaxis

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# Introduction

- **Idiopathic Anaphylaxis (IA): A syndrome of anaphylaxis with no recognized external stimulus**
- ❖ Clinically; urticaria or angioedema episodes with bronchospasm, hypotension, syncope, or gastrointestinal symptoms with or without upper airway compromise, with infrequent or frequent episodes (<6/yr or >6/yr)
- **It is estimated to affect 30,000 people in the US**
- **Initially described in adults and first reported in 1978, it has subsequently been reported in children**
- **Malignant IA (MIA): IA that cannot be controlled on <30 mg of prednisone daily or <60 mg of prednisone on alternate days** (Ditto et al. ; J. Allergy Clin Immunol 1997;100 : 320-326)

# Diagnosis of IA

- A diagnosis of exclusion
- A thorough history and physical examination
  - ❖ food and additive ingestion, medications, and exercise
- Specific IgE antibodies
- Controlled challenge procedures

# Differential Dx. of IA

- Vasovagal (neurocardiogenic) syncope
- Syndromes associated with flushing e.g. metastatic carcinoid, pheochromocytoma
- Systemic mastocytosis
- Hereditary / acquired angioedema
- Postprandial syndromes e.g., scromboid poisoning, food-induced anaphylaxis
- Psychiatric disorders such as panic attacks or vocal cord dysfunction
- Other causes of shock and cardiovascular or respiratory events

# Therapy of IA

- **Corticosteroid-independent IA**
  - ❖ Hydroxyzine, albuterol and epinephrine
- **Corticosteroid-dependent IA**
  - ❖ Prednisone
  - ❖ Ketotifen
  - ❖ **Omalizumab?**
- **Malignant IA (MIA)**
  - ❖ **Omalizumab?**

# Aim

- **We report a trial of Omalizumab in a steroid-dependent patient with MIA**
- **Discuss the importance of Omalizumab as a steroid sparing agent in the treatment**

# Patient-1

- 20-year-old AA female with anaphylactic attacks since 14 years of age
- Attacks begin with the precedent skin tightness, hives and hot feeling without burning followed by swelling on arms/ legs/ hands and sometimes progressing to throat swelling
- Attacks averted by taking 60 mg Prednisone or Epinephrine prn.
- Although she outgrew asthma and avoided from tomato and seafood, she has been having these attacks frequently twice-thrice /week
- OCP did not affect her symptoms.
- In the PMH, she had chronic rhinitis, severe persistent asthma and allergies to house dust, tomatoes and seafood
- **Family history was noncontributory**

# Patient-11

- When diagnosed with IA and she was put on a `Patterson regiment` including Prednisone (1-2 mg/kg/d) + Albuterol (2 mg tid) + Hydroxyzine (25 mg tid) with a great benefit.
- However, when we were able to taper Prednisone dose down to 5 mg on every alternate day, the episodes reappeared.
- Attacks that could not be controlled on <30 mg of prednisone daily and MIA diagnosis was made

# Labs - I

- CBC and ESR were normal.
- Ig G, A, and M were normal. IgG1 and IgG3 were slightly low.
- Ig E was elevated (408 IU/ml).
- Specific IgE was positive for Crab (0.43).
- Lymphs. subpopulation analysis was normal. Although CD4<sup>+</sup>/CD25<sup>+</sup> - cell numbers were increased, CD3<sup>+</sup>/HLA-DR<sup>+</sup> - cell numbers were low.

# Labs -II

- C1 esterase function/antigen levels, C1q, C2, C3, C4 and CH50 were all normal
- During an episode, tryptase and histamine levels were normal
- Urinary 5-hydroxyindolacetic acid, methylhistamine, and catecholamines were normal

# Labs - III

- TSH, T3 uptake, free-T4 and T4 were normal.
- ANA, dsDNA, RNP, SS-A, SS-B, smith antibody, and anti-thyroid auto antibodies were negative.
- Hepatitis B virus DNA and Hepatitis C RNA were negative.
- Punch (skin) biopsy revealed perivascular neutrophilic and eosinophilic collagenosis, compatible with urticaria and urticarial vasculitis.

# Result

- After being steroid-dependent for 4 years, Omalizumab, 225 mg sc every 2 weeks was started
- Omalizumab injections decreased her attack frequency and severity as well as the prednisone dose

# Conclusion

*Omalizumab may be worth to try in this desperate syndrome especially when the patient has additional allergic disorders*